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ABSTRACT

Designed to acquaint teachers, administrators and other school personnel with phenylketonuria (PKU - an inborn error of metabolism which requires dietary intervention), the booklet reviews school problems related to the condition. Introductory information concerns the nature, treatment, and screening and diagnosis of PKU. Diet management is discussed, and the role of school personnel in maintaining the diet is explained. The parents' part in informing the school of their child's condition is stressed. Briefly touched on are the PKU child's emotional needs to be treated as much like non-PKU children as possible, and the finding that treated PKU children have no different educational needs compared to non-PKU children. A resource section lists eight related written materials. (CL)

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PKU AND THE SCHOOLS

INFORMATION for TEACHERS, ADMINISTRATORS and OTHER SCHOOL PERSONNEL

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Special Terms

Two abbreviations are used which may be unfamiliar to the reader: *PKU*, the short version for phenylketonuria, and *phe*, which stands for phenylalanine—the amino acid which the child with PKU cannot metabolize. Both terms are common abbreviations, and parents and children with PKU will use them meaningfully.

Preface

The purpose of this booklet is to acquaint teachers, administrators and other school personnel with the current understanding of phenylketonuria (PKU), and to identify the psycho-educational implications of the condition. It provides suggestions to enable schools to educate children with PKU in accordance with the Doctrine of the Least Restrictive Environment, as mandated by Public Law 94-142. More extensive references to treatment of PKU, and sources of literature for parents and the PKU child are also provided.

The publication is based primarily on the experiences developed within the Collaborative Study of Children Treated for Phenylketonuria. This project which was started in 1965 is a joint effort of 15 clinical programs coordinated through Children's Hospital of Los Angeles and supported by the Bureau of Community Health Services through funds appropriated under title V of the Social Security Act (Grant No. MCT-000466).

Participants in the PKU Collaborative Project include:

Children's Hospital
Los Angeles, California

Regional Center for the
Developmentally Disabled
Childrens Hospital and Health
Center
San Diego, California

University of Colorado School of
Medicine
Denver, Colorado

Mailman Center
University of Miami
Miami, Florida

Cook County Hospital
Chicago, Illinois

Children's Memorial Hospital
Chicago, Illinois

Child Development Clinic
University of Iowa
Iowa City, Iowa

University of Maryland School of
Medicine
Baltimore, Maryland

Johns Hopkins Hospital
Baltimore, Maryland

Children's Rehabilitation Center
Children's Hospital
Buffalo, New York

State University of New York
Syracuse, New York

Children's Medical Center
Tulsa, Oklahoma

University of Texas Medical School
Galveston, Texas

University of Washington
Seattle, Washington

Waisman Center on Mental Retardation and Human Development
University of Wisconsin
Madison, Wisconsin

Introduction

Phenylketonuria, called PKU for short, is an inherited condition which prevents the child with this disorder from normally metabolizing—or using—phenylalanine, one of the essential amino acids found in all protein foods. Unless the condition is detected and treatment initiated within the first few weeks after birth, this hereditary biochemical abnormality will prevent normal brain development and usually results in severe mental retardation. Other manifestations such as skin rash, excessive restlessness, irritable behavior and a pungent, musty body odor may also appear.

Fortunately, detection of the condition shortly after birth through the use of a routine screening procedure has become standard practice in most States. Institution of a phenylalanine-restricted diet during the first 30 days of life appears to be effective in preventing the damaging effects of PKU. Since treatment requires almost entire elimination of foods high in protein from the PKU child's diet, special care must be taken to maintain enough, but not too much, phenylalanine in the child's system. Such a balance is obtained only through careful nutrition and medical supervision, and considerable effort by the parents.

Additional problems in maintaining the diet are encountered when the child with PKU enrolls in school. All children like to "swap" items from their lunches, and there may be considerable peer pressure for children with PKU to avoid special foods so as not to appear different to their schoolmates. Educators should be familiar with PKU in order to be supportive to the child and the parents, and to assist with diet management during school hours.

While the effectiveness of diet treatment has been adequately established, it is not yet clear when it is safe to discontinue or relax the diet restriction of phenylalanine. A study to obtain this information is currently underway as part of a national PKU Collaborative Study, and teachers in schools enrolling children from this study population have assisted in data collection.

The Benefits of Treatment

Untreated or late-treated children with PKU develop severe mental retardation. Therefore, it is imperative that the diagnosis of PKU be established within the neonatal period (first 30 days of life) before the onset of mental damage. In 1979, 46 States had blood screening programs to detect PKU shortly after birth, and similar programs are being developed in other countries throughout the world.

When treatment is instituted within the first few days of life, and parents are able to provide the child with a properly prescribed diet, normal development can be anticipated. A few years ago there was some disagreement expressed in the medical literature concerning the value of the phenylalanine-restricted diet, and some clinical opinion that the diet was detrimental to physical growth and development. However, there is now abundant evidence which validates the effectiveness of the special diet in

preventing mental retardation. Admission of PKU individuals to State institutions serving the mentally retarded has almost been eliminated (MacCready, 1974). At this time, most treated PKU children are enrolled in public schools. They usually cannot be distinguished from normal children, except by their restriction to a special diet.

Screening and Diagnosis

From 1934, when PKU was discovered by Dr. Asborn Folling in Norway, through the 1950's, the condition was detected with a urine "wet diaper" test. This method has many disadvantages, and since 1964 has been discontinued in favor of a blood test which should be administered just before the baby leaves the hospital. If performed earlier than four days of life, the test should be repeated by four weeks of age (Guthrie & Susi, 1963). When the baby has higher than normal levels of phenylalanine in the blood, confirmatory tests are performed, and if the diagnosis is established, diet treatment begins as soon as possible. Results from the PKU Collaborative Study clearly indicate a loss in later IQ scores if the baby is not put on the diet within the first 30 days of life.

Diet Management

The fact that PKU involves a single amino acid—phenylalanine (phe)—might lead one to believe that the diet is thus easy to prepare and manage. On the contrary, phe is an essential building block of the children's protein intake. Thus, the children must have a minimum amount of phe in the foods eaten each day, but the phe in the blood should not exceed about 10 mg%. (The normal level is between 1 and 3 mg%.) A normal diet is high in protein foods: milk, meat, fish, eggs and nuts. All of these must be eliminated or provided in prescribed amounts. Fruits, vegetables and cereals contain smaller amounts of phe, and can be eaten by the PKU child providing the total amount of phe ingested in a given day is not excessive. This delicate balance is obtained by careful measuring of all food and calculating the phe intake on a daily basis. As the children grow older, increase in weight, and new foods are added to their diet, the amount of each kind of food must be adjusted accordingly.

In order to supply the other amino acids required for normal growth and development, special formulas such as LOFENALAC® * have been prepared. Most school-aged children will be on a similar formula (Product 8229) also produced by Mead-Johnson. These formulas contain all the amino acids except that most or all of the phe has been removed. They also have been fortified with fats, carbohydrates, vitamins and minerals, and thus provide a balanced diet base. Dietitians, nutritionists, and parents have devised extensive cookbooks of recipes using one of the special formulas to produce breads, cookies, puddings, "ice cream," and other foods. (See Annotated Bibliography.)

*LOFENALAC® is a registered trademark of the Mead-Johnson Laboratories of Evansville, Indiana.

Special diet management problems may occur at school. The child with PKU, for example, cannot have the usual "snack time" carton of milk. Special events, such as a birthday, may involve a cake containing forbidden high-phe food products. Because the PKU child should not eat these foods, he/she is identified as different. Teachers and administrators must be aware of potential problems and take preventive measures to insure that the PKU child can maximally participate in all activities, including snack time and parties, by having suitable alternative foods available.

If a school lunch program is provided, many of the foods can be safely eaten by the PKU child. It is often helpful to send the week's menu home so that the parents can indicate which items and how much the child can have and which must be avoided, and return the menu to the teacher. Older children will often be able to identify those foods which are on their diets themselves. The ability of PKU children to participate in the same lunchroom program as their classmates will lessen the feelings of being different, and thus permit healthy social and emotional growth.

Role of School Personnel

As a general rule, children who have been properly treated for PKU are normal, healthy youngsters, subject to the same problems, likes and dislikes of other school children of the same age. Variations in height, weight or other physical aspects of a given PKU child are probably unrelated to the existence of PKU, or to diet treatment. The PKU Collaborative Study found that children who have been satisfactorily treated for PKU have normal growth and development. Likewise, the IQ scores of the children at 4 and 6 years of age were very near the national mean score of 100 (Dobson et al., 1977). An evaluation of psycho-educational measures, such as the Illinois Test of Psycholinguistic Abilities, the Frostig Visual Perception Test and the Bender-Gestalt Test, revealed no special profiles of strengths or weaknesses in children with PKU. As a group, those PKU children who had entered school were achieving at or above the national norms, as measured by the Wide Range Achievement Test.

Individual differences will be found, of course, as they will in any group of children. It is important to remember that such differences are most likely unrelated to the existence of PKU or to the special diet. Thus, if special academic or behavioral problems develop in a student with PKU, the usual referral processes should be used.

The age at which diet can be discontinued safely is presently not established, and clinics differ in the age at which a free diet is instituted. It is anticipated that some children may manifest behavioral symptoms associated with changes in diet restrictions which lead to elevated phenylalanine blood levels. In some children this may result in reduced attention span or hyperactivity. The PKU Collaborative Study is currently investigating this problem, and school personnel may be asked to assist in providing data concerning a PKU child who is either 1) continuing the special low-phe diet, or 2) gradually increasing the amount of phe until a normal diet is achieved. If the child requires special educational treatment during the collection of data, the PKU clinic should be informed.

Circumstances may warrant a direct educational program by the teacher or other school personnel to explain the child's need for a special diet to other children in the class, and perhaps to their parents. Young children have an enormous curiosity, and can be extremely cruel to a classmate whom they discover is different in some way. If this occurs, the teacher should consult with the parents and with the PKU Clinic to consider an educational program about PKU for all children in the class. This could be approached through a "PKU cooking project" where all children in the class prepare and taste a special PKU food, such as low-phe cookies. Recipes for cookies and other PKU foods are found in "The PKU Cookbook" published by the University of New Mexico (Read et al., 1976), and "Low Protein Cookery for PKU" by Virginia E. Schuett (University of Wisconsin Press, 1977).

Other audio-visual and printed materials designed for use with parents and children are also identified in the annotated bibliography.

The PKU Child at School

By the time PKU children reach school, their parents will have had several years' experience with dietary management and related problems. The PKU child's medical experiences will be different from other siblings due to the special diet and frequent visits to the PKU clinic for blood tests and other examinations. Therefore, the schools must be sensitive not only to the child's nutrient needs but also to his emotional and social needs.

PKU clinics have assisted parents in "educating" the educators about their PKU child's diet problems. A sample letter from a clinic to parents reads as follows:

Dear Teacher:

_____Name_____ has PKU and is on a special diet because of this. Here is an explanation to help you understand what it is and how to deal with it.

PKU, which is short for phenylketonuria, is an inherited defect in body metabolism. Children with PKU lack an enzyme necessary to metabolize or break down phenylalanine, an amino acid (protein) found in most foods. As a result, phenylalanine is not completely used in a normal manner in the body, and it collects in the body tissues and blood. The excess phenylalanine acts to prevent proper physical development and mental growth of the child.

PKU, if diagnosed early as _____Name_____'s was, can be successfully treated by dietary regulation of phenylalanine. By keeping the dietary phenylalanine low, it is possible to keep the level in the blood at a normal level. Hence, mental and physical development can proceed at a healthy normal rate.

Some phenylalanine is necessary for growth in the amount of the PKU diet. Consuming more phenylalanine than is necessary for growth will result in brain damage. Therefore, a person with phenylketonuria cannot consume any protein except for a low phenylalanine formula. High protein foods such as meat, fish, milk, eggs, cheese, most breads and cereals must be avoided completely. Many foods may be eaten freely (free foods).

_____Name_____ is beginning to be very aware of his/her diet allowances and restrictions. He/She needs your assistance to continue learning about it. Please do not feed him/her anything that is not on this list or approved by his/her parents. The diet is very important and a loose approach to it will be very detrimental to _____Name_____'s health.

Free Foods	Foods To Avoid	Foods That Can Be Eaten In Limited Quantities
Apple juice	Meat	Any fruit (canned or fresh)
Apple cider	Eggs	Vegetables (cooked or raw)
Hard candy	Fish	Marshmallows
Jelly beans	Milk	Pretzels
Lollipops	Cheese	Vanilla wafers
Soda pop	Chocolate	Rice Krispie Treats
Kool-aid	Nuts	Popcorn
Lemonade	Peanut butter	Animal crackers
Popsicles	Ice Cream	Assorted baked goods (cookies, cakes, bread) *sent from home*
Tang	Yogurt	Saltines
Apples		Ritz Crackers
Jelly		Wheat Thins
Applesauce		Waverly Wafers
		Graham crackers
		Fruit or vegetable juices
		Potato chips (10 or less)

If a food experience will be a part of a future lesson plan or if special treats are being brought by another child, please notify me ahead of time, if possible. That way, I can plan something similar or appropriate for _____ within his diet limits.

Thank you very much for your cooperation.

The PKU child's emotional needs are best met by treating him/her as much like non-PKU children as possible. All children require support systems at school and at home to develop their self-concept. Children with special needs often need sensitive support from parents and teachers to feel accepted and "normal."

Experience with PKU children in preschool and elementary school programs has provided answers to many questions:

—While children are curious about each other and are quick to note differences such as a special diet, they also are able to understand the concept of a "food allergy." Thus, no attempt should be made to hide the fact that a PKU child's lunch is different, nor should any lengthy explanation be required.

—Since many schools have the practice of providing a "lunch supervisor" to permit teachers a period away from their children to have their own lunch, the problem of preventing "swapping" food items might seem unmanageable. Probably even more important for the schools to remember is the need *not* to draw excessive attention to the PKU child's differences. Occasional indiscretions are not critical, and as children grow older they must take more and more responsibility for their own dietary management. Thus, while the PKU child should be observed unobtrusively, the only action needed by the teacher or lunchroom supervisor is to notify the parent when any high-phe foods have been eaten. The parent may need to adjust the child's intake to maintain proper intake during this period. The parent may also want to work with school personnel to prevent recurrence of the difficulty, and to supplement the instructions given the PKU child.

—There is a tendency to overreact to the diagnosis of PKU by some school personnel. One school classified a PKU child as "physically handicapped" and authorized a limited, adaptive physical education program. This is totally wrong: the PKU child is different from other children only in terms of restricted food choices, not in activity or general health. Similarly, should a PKU child become ill at school, the regular school procedures should be followed. PKU is not associated with any sudden, dramatic episodes such as the insulin reaction experienced by diabetics, or the grand mal seizure of an epileptic. Thus, any symptoms of illness will be unrelated to the child's PKU.

—Similarly, the PKU child's educational needs are unaffected by the PKU. The National Collaborative Study has found no differences between treated PKU children and their normal siblings on a number of school-related measures, such as visual perception, psycho-linguistic profile, IQ and achievement test scores. Thus, the PKU child needs to be treated as any normal child, with strengths and weaknesses, likes and dislikes, and a learning style unrelated to the PKU condition. A child with PKU who has been well managed with diet should not require special programming, but should be on an age-appropriate developmental level.

—At times educators become particularly concerned when the curriculum calls for nutrition or food classes. The basic rule for the schools to follow is to establish communication with the parents. They will have, or know of, materials (See Annotated Bibliography) useful to supplement the regular school texts, thus enabling the PKU child to be included in such classes. In all such programs the stress should be on the necessity for the individual to make choices, and thus the PKU child becomes part of the lesson, and is not pointed out as different. The need for PKU children to monitor their intake of food can serve as a model, especially for children with other kinds of diet problems.

— In the final analysis, the teacher of a child with PKU has just two major responsibilities:

(1) **To treat the child with PKU as a normal, healthy member of the class, no different from anyone else in terms of social, emotional, physical and academic growth and development.**

(2) **To establish and maintain open communication with the PKU child's parents so as to reinforce parent teaching and monitoring of food eaten at school.**

Resources

For additional information or assistance beyond that presented in this booklet and the references provided, the following sources are suggested:

(1) PKU clinics. Names and addresses of major PKU clinics in the United States are available from the Office for Maternal and Child Health, Bureau of Community Health Services, 5600 Fisher Lane, Rockville, Md. 20857.

(2) State Departments of Public Health. Most States have developed screening programs for PKU and other inborn errors of metabolism, and have educational materials and directories of available resources for parents and teachers.

Annotated Bibliography

To help you learn more about PKU and the needs of the child with PKU, special materials have been written by parents and health care professionals. Most of the materials listed below are available through the staff at a PKU clinic. Brief comments have been added to help you decide which item will be most useful to you at a specific time in the PKU child's development.

Living with PKU

PKU Clinic staff and parents. University of Colorado Medical Center, 4200 East 9th Avenue, Box 2741, Denver, Colorado 80220. 12 pages. Distributed by Mead Johnson Laboratories, Evansville, Indiana 47721. Revised, May 1978. Single copies free on request.

Helpful information and reassurance to parents of PKU children. Using a question and answer format, the brochure deals with the most frequent concerns of parents. Parents have written from their own personal experience.

Diet Management of PKU for Infants and Preschool Children

Acosta, Phyllis B., and Wenz, Elizabeth. U.S. Department of Health, Education, and Welfare, Public Health Service, Health Services Administration, Bureau of Community Health Service, Rockville, Maryland 20857. DHEW Publication No. (HSA) 77-5209, 1977. Single copies free on request.

A readable and comprehensive discussion of behavior management of PKU; includes food lists.

What is PKU?

Iris Crump. San Diego Regional Center for the Developmentally Disabled, Childrens Hospital and Health Center, 8001 Frost Street, San Diego, California 92123, 1975. 14 pages. Single copies free on request.

A simplified explanation of the condition for affected families.

PKU: A Diet Guide for Parents of Children with Phenylketonuria

Phyllis Acosta, Elizabeth Wenz, Graciela Shaeffler, and Richard Koch. Division of Child Development of the Los Angeles Childrens Hospital, Los Angeles, California. 1969, revised 1974. 20 pages. Distributed by Mead Johnson Laboratories, Evansville, Indiana 47721.

Basic description of diet for PKU. Includes instructions on formula mixing, lists foods permitted, and deals with food related concerns.

PHE and Me

MLK Film Products, 837 Jennifer Street, Madison, Wisconsin 53703. 8 mm, color, 7 minutes. Available on request.

A cartoon film about phenylalanine and its use in the body. Discusses why some children can get too much phe and how to know the right amount of phe for the child. Best for 5-year-olds and over.

The PKU Cookbook

Elizabeth Read, Elizabeth Wenz, Roslyn A. Duffy, Nancy S. Wellman, Aissa Acosta, and Phyllis B. Acosta. University of New Mexico Printing Plant, Albuquerque, New Mexico 87101. Available from Phyllis B. Acosta, School of Medicine, Emory University, Atlanta, Georgia 30322. Single Copies $10.92.

Low Protein Cookery for Phenylketonuria

Virginia E. Schuett. Waisman Center on Mental Retardation and Human Development, 1500 Highland Avenue, Madison, Wisconsin 53706, 1977. 374 pages. Distributed free to families of children with PKU through PKU treatment centers.

Comprehensive compilations of recipes for low-phe foods. Many of the recipes could be incorporated into classroom cooking experiences.

You and PKU

Margaret Taylor and Virginia E. Schuett, Waisman Center on Mental Retardation and Human Development, 1500 Highland Avenue, Madison, Wisconsin 53706, 1978. 44 pages. Distributed free of charge to families of children with PKU through PKU treatment centers.

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